Lateralizing features by EEG were recorded in 12 (41%), and 7 had an MRI abnormality on the same side. (Cross JH et al. Early detection of abnormalities in partial epilepsy using magnetic resonance. Arch Dis Child July 1993;69:104-109). (Respond: Dr Helen Cross, Neurosciences Unit, The Wolfson Centre, Mecklenburgh Square, London WC1N 2AP, UK).

COMMENT. An MRI finding of hippocampal sclerosis was documented as early as 4 years of age in one patient in this study, and progressive hippocampal abnormalities over a six month period were apparent in one other. The MRI as employed in this study can separate the causes and effects of seizures and help to clarify the relationship between early hippocampal damage in children and subsequent temporal lobe epilepsy.

Studies of hippocampal neuron loss and memory scores, before and after temporal lobe surgery for epilepsy at the Reed Neurological Research Center, University of California, LA, support a role for the left hippocampus in rote verbal memory. Patients with severe as compared with minimal left hippocampal neuron loss may be at risk for lower memory functioning postoperatively (Rausch R, Babb TL. Arch Neurol Aug 1993;50:812-817).

DEGENERATIVE DISEASES

LATE-ONSET FRIEDREICH'S ATAXIA

Three adult patients from one family with late-onset Friedreich's ataxia (LOFA) presenting after 25 years (mean age, 30 yrs) were compared with 13 children with classical FA presenting before 20 years (mean age, 13 yrs) and reported from the University of Tubingen, Germany, and St Mary's Hospital, London, England. Clinical presentation of LOFA and FA were similar, except that muscle wasting, foot deformity, and cardiomyopathy were absent in LOFA patients. Genetic linkage analysis using markers tightly linked to the FA locus on chromosome 9 showed that all affected members of the LOFA family, but not their unaffected siblings, had inherited identical paternal and maternal genotypes. LOFA may result from mutation within the FA locus, giving rise to a more benign and slowly progressive disorder. (Klockgether T, Chamberlain S et al. Late-onset Friedreich's ataxia. Molecular genetics, clinical neurophysiology, and magnetic resonance imaging. Arch Neurol Aug 1993;50803-806). (Reprints: Dr Klockgether, Department of Neurology, University of Tubingen, Hoppe-Seyler-Str 3, W-7400 Tubingen, Germany).

COMMENT. With the exception of age of onset, all patients with LOFA satisfied the basic diagnostic criteria for classical Friedreich's ataxia: 1) progressive ataxia; 2) family history with autosomal-recessive

inheritance; 3) loss of tendon reflexes in lower limbs; 4) dysarthria; and 5) posterior column signs. The more benign 'Acadian' subtype (Barbeau et al, 1984) has previously been differentiated from the classical, French and French Canadian, type of FA and has been attributed to a mutation at the same locus (Keats BJ, Chamberlain S et al. Am J Med Genet 1989;33:266).

LORENZO OIL THERAPY FOR ADRENOLEUKODYSTROPHY

Dietary therapy with glycerol trioleate and glycerol trierucate (Lorenzo oil) was tested in 108 adult patients with adrenomyeloneuropathy phenotype of adrenoleukodystrophy (ALD) at Johns Hopkins Hospital and the Kennedy Krieger Institute, Baltimore, MD. Pattern-reversal visual evoked potentials were used to evaluate visual pathways before and after treatment for 1 year. Very-long-chain fatty acid (VLCFA) levels were markedly reduced, but visual evoked potentials remained abnormal or became abnormal. No patients improved, and there was no evidence that reduction in VLCFA levels benefited or retarded demyelination of visual pathways. (Kaplan PW, Moser HW et al. Visual evoked potentials in adrenoleukodystrophy: a trial with glycerol trioleate and Lorenzo oil. Ann Neurol Aug 1993;34:169-174). (Respond: Dr Kaplan, Department of Neurology, Francis Scott Key Medical Center, 4940 Eastern Avenue, Baltimore, MD 21224).

COMMENT. Moser, in an editorial (Ann Neurol Aug 1993;34:121-122), reviews the development of therapies for adrenoleukodystrophy and describes Lorenzo oil as "a prematurely amplified hope." Trials of the oil in Europe and the USA have failed to demonstrate a significant effect on the rate of progression of the childhood cerebral form of ALD. A possible preventive effect of Lorenzo oil in patients who have not yet developed the neurological disability is under investigation. Of 61 asymptomatic patients treated for a few months up to 4 years at the Kennedy Krieger Institute, one has developed childhood cerebral ALD and 7 have shown progressive demyelination on the MRI. Longer follow-up is required to determine the outcome in the 53 who remain asymptomatic. A depression in platelets was an unexpected side effect of Lorenzo oil therapy.

METACHROMATIC LEUKODYSTROPHY VARIANTS

Clinical, pathological, imaging, and genetic findings in a family with multiple allelic mutations of metachromatic leukodystrophy (MLD) are reported from McGill University, Montreal, and McMaster University, Hamilton, Canada. The propositus, a 23-year-old man, presented at age 18 with a 3-year history of clumsiness and stiffness of gait. Two maternal uncles, ages 48 and 56, were also neurologically impaired. Two siblings, a brother aged 30